

Eagle's Syndrome: A case report of bilateral elongated styloid process

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Abstract

Eagle's syndrome or Elongated styloid process syndrome is an uncommon condition which presents with recurrent orofacial pain, globus sensation in the throat, referred otalgia and dysphagia owing to elongated styloid process or ossified stylohyoid ligament. There is a myriad of related pain syndromes in this region making the diagnosis challenging. Therefore diagnosis can be made only through elaborate history, careful clinical examination and confirmed with elongated styloid process or calcified stylohyoid ligament. Here we are discussing about a case with chief complaint of facial pain in a 53 year old female, which was diagnosed to have bilateral elongated styloid process.

Keywords: Eagle's syndrome, elongated styloid process, orofacial pain.

INTRODUCTION

The styloid process is a bilateral, pointed bony projection arising from the temporal bone that is situated at the front of the stylomastoid foramen. The normal length ranges from 2cm to 3cm and it is considered elongated when it is longer than 3cm¹. In 1937, Eagle described a syndrome characterized by an elongated styloid process and pain in the cervicofacial region. The prevalence is about 4% and most of them being asymptomatic⁴ and between 4% and 10% of the patients having an elongated styloid process experience the symptoms⁷. There is a female predilection and average age of symptomatic patient is usually 40 years. This is attributed to the fact that as age advances, the elasticity of the soft tissues and the associated ligaments is lost, putting increased pressure on the adjoining hard tissues.

The presenting symptoms include dull, aching pain on either side of the throat, difficulty in swallowing, foreign body sensation in the throat, pain in the facial region, recurrent headache and vertigo. Since these symptoms mimic many maxillofacial and oropharyngeal disorders and neuralgias, a thorough clinical history, examination, and radiological assessment are necessary for attaining a diagnosis. Here, we present one such case of Eagle's syndrome in a 53 year old female patient.

CASE REPORT

A 53 year old female patient reported to the

Department of Oral Medicine and Radiology with a complaint of intermittent dull aching pain in the left side of face and neck region since 3 weeks. Pain is maximum during morning and gradually reduces during daytime and associated with headache, difficulty in drinking water, loss of balance and diminishing vision of left eye. Her medical history reveals osteoarthritis of joints. There was neither history of trauma or previous surgery nor any identifiable trigger points. On inspection of TMJ there was normal mouth opening and pain was present on maximum mouth opening. There was no deviation, no deflection and no restriction in lateral or protrusive jaw movements. On palpation there was tenderness on maximum mouth opening. Based on the history and clinical examination provisional diagnosis was given as Myofacial Pain with TMJ arthralgia.

As a part of investigation an orthopantomograph (Figure 1) and double TMJ view (Figure 2) was taken which revealed bilateral elongated segmented styloid process measuring approximately 5cm on the right side and 5.5cm on the left side. Based on the radiographic findings, we came to a final diagnosis of bilateral Eagle's syndrome.

DISCUSSION

Eagle's syndrome also referred as Elongated styloid syndrome is sequelae of either an elongated styloid process or mineralisation of stylohyoid complex. It causes vague orofacial pain or pain along the distribution of external



Figure 1. Orthopantomograph shows bilateral elongated segmented styloid process measuring approximately 5cm on the right side and 5.5cm on the left side.



Figure 2. Double TMJ View shows bilateral elongated segmented styloid process was noticed measuring approximately 5cm on the right side and 5.5cm on the left side.

and internal carotid arteries, dysphagia, sore throat, glossodynia, otalgia, vertigo and headache. The most consistent symptom probably is the pharyngeal pain.

Approximately 4% of the population has elongated styloid process⁴ but only 4 to 10.3% of this group is symptomatic⁷. There is a female predilection and the average age of presenting symptoms is around 40 years. The incidence of mineralised and elongated styloid process (MESP) is reported as 1.4% to 30%. But majority of this population remains asymptomatic and few become symptomatic as age advances. This is attributed to the fact that aging results in loss of elasticity of the soft tissues and the associated ligaments putting increased pressure on the adjoining hard tissues.

An ossifying process of stylohyoid ligament was first described by Pietro Marchetti in 1652². Later in 1937 a syndrome characterized by an elongated styloid process and pain in cervicofacial region was first described by

Dr. Eagle³. Eagle reported around 200 cases in 20 years and concluded the normal length of styloid process is between 2.5cm to 3cm. He also noted that even a minor medial deviation of the styloid process can lead to recurrent severe atypical facial pain⁵. He classified the syndrome as Carotid artery type and Classic type. The Classic type is followed by tonsillectomy when the scar tissue underneath the tonsillar fossa compresses the V, VII, IX and X cranial nerves, causing discomfort and difficulty in swallowing and foreign body sensation in the throat. The Carotid artery type presents as headache and nerve problem due to the inflammation of sympathetic nerve plexus. The above described case is of the carotid artery type.

Langlais et al. has given a radiographic classification of the elongated and mineralized stylohyoid ligament complex⁸. This classification includes three types of abnormal radiographic appearances and four patterns of calcification/mineralization (see Tables 1 and 2).

Table 1 : Types of elongation of styloid process

| Types | Characteristics |
|--------------------|--|
| Elongated | Uninterrupted elongation(>25-28mm) |
| Pseudo articulated | Less frequent than elongation; the styloid process is joined to the mineralized stylomandibular or stylohyoid ligament by a single pseudo-articulation, usually located superior to the inferior border of the mandible. |
| Segmented | Short or long noncontinuous portions of the styloid process or interrupted segments of mineralized ligament |

Table 2 : Patterns of calcification

| Patterns | Characteristics |
|----------------------|---|
| Calcified outline | Appearance of a long bone with a thin radiopaque cortex and a central radiolucency that constitutes most of the process. |
| Partially calcified | Thicker radiopaque outline, with almost complete opacification and small and occasionally discontinuous radiolucent core. |
| Nodular complex | Scalloped outline and may be partially or completely calcified with varying degrees of central radiolucency. |
| Completely calcified | Totally radiopaque with no evidence of a radiolucent inner core. |

Treatment for Eagle's syndrome can be either conservative or surgical. Conservative treatment is by non-steroidal anti-inflammatory drugs and injection of corticosteroids and local anaesthetics into the tonsillar fossa. Surgical excision by the intraoral or extraoral approach has often been successful⁶.

CONCLUSION

Eagle's syndrome should be included in the list of differential diagnosis for evaluating headaches, tonsillar discomfort, pain referred to the jaw region, dysphagia and difficulty to perform lateral jaw movements. The presenting symptoms differ from person to person and there is a multitude of disease with similar symptomatology that makes the diagnosis challenging. Therefore it is very much important to know in detail about all the possible presentations of this rare clinical condition.

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